Schwannoma of the External Auditory Canal: An Exceptional Location

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Solitary schwannoma is a benign, encapsulated tumour of Schwann cell origin, therefore the olfactory and optic nerves are never affected. About 25 %-45 % of all schwannomas occur in the head and neck but schwannoma of the external auditory canal is a rare finding, and we have found only 6 previous cases reported in the literature world-wide. Our patient was discovered by chance during a stapedectomy because the tumour was sited in the external auditory canal without distorting it.

Key words: Schwannoma of external auditory canal. Benign encapsulated tumour. Excepcional location.

Schwannoma de conducto auditivo externo: una localización excepcional
El schwannoma es un tumor benigno encapsulado que puede afectar a cualquier nervio del organismo que tenga células de Schwann, nunca al nervio olfatorio ni el óptico. Un 25-45 % aparece en cabeza y cuello, pero en el conducto auditivo externo es excepcional, y solamente se han publicado en la literatura mundial otros 6 casos previos. Presentamos un caso descubierto accidentalmente al realizar la incisión en un conducto normal, no abombado, durante una estapedectomía.

Palabras clave: Schwannoma de conducto auditivo externo. Tumor benigno encapsulado. Localización excepcional.

INTRODUCTION

A solitary schwannoma, also referred to as neurinoma or neurilemmoma, is a well-encapsulated benign tumour, moderately painful and slow-growing, that originates in the Schwann cells of the nerves, therefore any nerve in the body can be the source of these tumours, except for the optical and olfactory nerves, which do not have any of these coating cells.[1] Around 25%-45% of all schwannomas appear in the head and neck,[1-3] with the 8th cranial pair the most frequent intracranial location and the side of the neck the most frequent extracranial location.[1-4]

Schwannomas of the external auditory canal (EAC) are very infrequent, and we might even say exceptional, if we consider that there have only been 6 previous cases published in the world-wide medical literature.[3-8]

This paper presents a new case, the seventh reported in the world, that was found entirely by chance in the course of a stapedectomy, as the tumour did not protrude into the canal and appeared by surprise when effecting the incision on the EAC at the start of the surgical procedure.

CASE STUDY

A 55-year-old female diagnosed as having otosclerosis was subjected to a standard endoaural operation.

The patient had not reported any pain or discomfort in her ear and the auditory canal was absolutely normal during examination and did not present any tumouration or inflammation on the walls.

When the incision was made in the canal to raise the tympanomeatal flap, a whitish, well-defined spindle-shaped formation approximately 0.5 cm long appeared unexpectedly in the roof of the EAC and continued as a ribbon of similar characteristics towards the depths of the EAC. We were unable to interpret what it was at the time and it was sent to the pathology department in formol to be studied.

It was macroscopically described as an irregular fragment measuring 0.4×0.2 cm, brownish in colour and elastic consistency.

Under the microscope, the slices showed several circumscribed encapsulated nodules comprising interwoven bands of spindle-shaped cells of nerve origin (Figure 1). No

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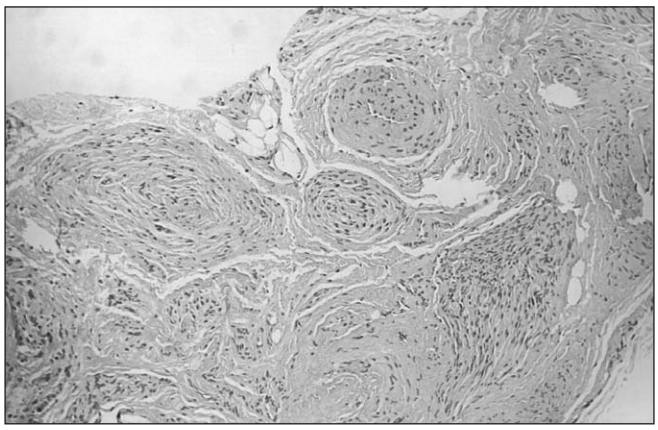

Figure 1. Encapsulated circumscribed nodes comprising a proliferation of spindle-shaped cells of nerve origin (HE, ×10).

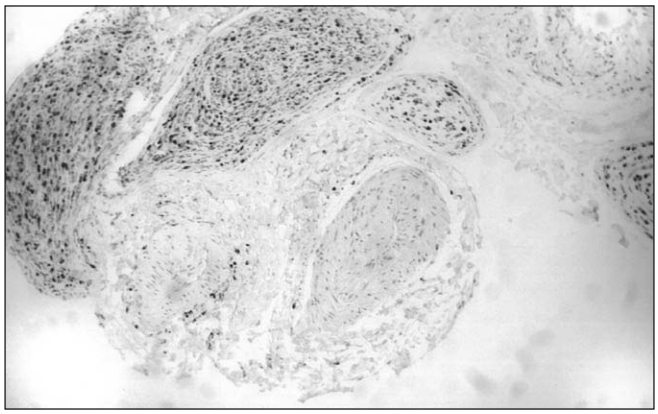

Figura 2. Immunohistochemical technique. Marked positive response for protein S-100 (×10).

mitosis or marked atypias were seen. Immunohistochemical techniques show a marked positive result for protein S-100, a marker for nerve-origin cells and negative values for AE1-AE3, a marker for epithelial cells, thus confirming the nerve origin of the lesion (Figure 2). Diagnosis: plexiform schwannoma.

DISCUSSION

Solitary schwannomas are benign tumours first described in 1908 by Verocay, who gave it the name of neurinoma; in 1974 Batsakis assigned it the name schwannoma,[5] although it has been known by several terms, such as neurinoma, neurilemmoma, mioschwannoma, schwannoglioma, etc.[4]

It affects any nerve in the body that has Schwann cells in the sheath, whether in the autonomous nervous system or in the peripheral system such as the cranial nerves. Since they do not have Schwann cells, the olfactory and optical nerves are not affected by this kind of tumour. Within the head, the 8th pair is the nerve most often affected, followed in order of frequency the sensory nerves as the motor nerves are only very rarely involved.[7]

The mean age of onset is 30-60 years and it is more common in women,[1] albeit this last finding is still controversial.[2]

Histologically, two growth patterns are distinguished in a single tumour, namely Antoni's types A and B,[1,4] with these two patterns alternating in very variable proportions.[1,4,7] Type A is characterized by poorly defined edges and a stroma that is rich in fibrillar material and long pale-nucleate cells in a palisade structure forming the so-called Verocay's bodies, whereas Type B has a looser structure with little cellularity. The protein S-100 marker for nerve tissue is intensely positive in schwannomas, especially in type A.

Since Cejas et al[5] published the first case of EAC schwannomas in 1988, only 6 cases have been reported in the world-wide literature prior to ours,[3-8] a statistic that excludes 2 cases published in Russian and located in the external auditory meatus[9,10] and another located on the tympanic membrane,[11] as these have been deemed not strictly the EAC.

In the EAC, these tumours are usually asymptomatic and paraesthesias or neuralgias are very infrequent; only when the tumour attains a sufficient size can it produce a sensation of blockage in the ear or cause cases of external otitis due to accumulation of debris or dampness in the bottom of the EAC.[2]

As the tumour arises out of the Schwann cells, it affects the nerve surface, unlike neurofibromas, born in the nerve fibres themselves, and therefore the nerve fibres would in principle only be squashed and not injured; this is a very important fact when considering surgery, as it is sometimes possible to identify perfectly the nerve where the tumour originates and when the tumour lies superficially across the nerve it can be removed without necessarily sacrificing the nerve.

Schwannomas or neurilemmomas rarely become malignant, but primary malignant tumours have been described, particularly in the neck.[7]

Treatment requires surgery following the endoaural route, as happened in our case, or from behind the ear if the tumour is too large,[3,7] and relapses are very infrequent.[1,7]

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